Case Report

Surgical Repair for a Coronary-Pulmonary Artery Fistula with a Saccular Aneurysm of the Coronary Artery

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The patient, a 69-year-old woman, had been diagnosed with a heart murmur. A chest X-ray at a local clinic had shown an abnormal shadow. Since CT revealed a 3-cm-diameter mass close to the pulmonary artery, we performed a coronary angiography and diagnosed her as having a coronary artery aneurysm associated with a coronary-pulmonary artery fistula. We incised the aneurysm under cardiac arrest, the wall of which had three openings that were suture closed from the inside and outside. The coronary-pulmonary artery fistula was suture closed. A postoperative angiography confirmed the disappearance of the coronary artery aneurysm and the abnormal blood vessels. The patient had an uneventful postoperative course and was discharged on postoperative day 15. We report a rare case of coronary-pulmonary artery fistula with a coronary artery aneurysm for which surgery was followed by an uneventful postoperative course. (Ann Thorac Cardiovasc Surg 2009; 15: 194–197)

Key words: coronary artery, coronary artery aneurysm, coronary-pulmonary artery fistula

Introduction

A coronary-pulmonary artery fistula is an occasionally observed congenital anomaly; it is rarely associated with a coronary artery aneurysm. It causes few symptoms and is often detected incidentally by abnormal chest X-ray shadows or coronary angiography. Because studies have reported that even small coronary artery aneurysms about 1 cm in diameter can rupture, it is important to consider surgical indications and timing.

Case Report

The patient was a 69-year-old woman (height: 159 cm; weight: 75 kg; blood pressure: 120/68 mmHg; pulse: 70/min). She was being treated for hypertension and hypothyroidism. A chest X-ray showed prominence of the left atrial appendage, no pulmonary congestion, and a cardiothoracic ratio of 49%. A chest X-ray also showed an abnormal shadow, and contrast CT revealed a 3-cm-diameter mass close to the pulmonary artery (Fig. 1). A coronary angiography led to the diagnosis of a coronary-pulmonary artery fistula with a coronary artery aneurysm. The fistula was formed between the anterior descending branch of the left coronary artery and the pulmonary artery. A right coronary-pulmonary artery fistula was also present, but no aneurysm formation was noted. On auscultation, a 2/6 Levine continuous murmur of maximum intensity was heard at the left sternal border at the second intercostal space. Although she had been followed with the diagnosis of patent ductus arteriosus (PDA), no PDA was present, and the heart murmur was considered to be due to the coronary-pulmonary artery aneurysm. Electrocardiography showed a normal sinus rhythm of 70 beats per min. Pressure measurements (mmHg) were as follows: pulmonary artery 34/12, pulmonary capillary wedge pressure (PCWP) mean 4, and aorta 158/79. TI^{201} stress myocardial scintigraphy revealed

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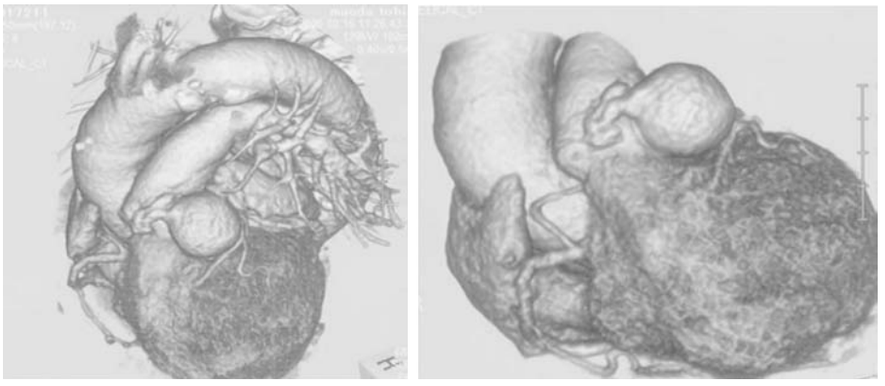

Fig. 1. CT image showing a 3-cm-diameter saccular coronary artery aneurysm close to the pulmonary artery.

no myocardial ischemia. Arterial blood gas sampling showed a very small amount of shunt flow (a pulmonary to systemic flow ratio [Qp/Qs = 1.02]). Cardiac ultrasound revealed good cardiac function: left ventricular ejection fraction (LVEF) = 73% and left ventricular dimension-diastole (LVDd)/Ds = 52/32 mm. There was no evidence of flow through the coronary-pulmonary artery fistula. The patient had no marked symptoms; however, since the aneurysm carried the risk of rupture, we performed surgery. Extracorporeal circulation was established by drawing blood from the superior and inferior venae cavae and draining it into the ascending aorta, and a cardioplegic solution was infused in an antegrade manner. The coronary artery aneurysm was incised under cardiac arrest. The aneurysm had a thin wall, was soft (Fig. 2), and contained no thrombus. Three blood vessels were seen to open into the aneurysm. The openings were suture closed from the inside of the aneurysm. Since the three inflow blood vessels communicating with the aneurysm were dilated, they were suture closed from the outside. A cardioplegic solution was infused to ascertain the absence of inflow into the aneurysm. The incised aneurysmal wall was closed with 4-0 polypropylene sutures using felt strips.

A histopathological examination showed that the aneurysmal wall was partially calcified and mainly composed of fibrotic tissue. Traces of the arterial wall, including the smooth muscle layer and elastic lamina, were scattered, suggesting that not only the external arterial wall and surrounding connective tissue, but also the aneurysmal wall underwent changes, mainly including fibrosis, because of the long-term persistence of the aneurysm. Postoperative CT and coronary angiography (Fig. 3) confirmed that the coronary artery aneurysm and abnormal blood vessels had disappeared, and blood

Fig. 2. Intraoperative photograph.

flowed through the anterior descending branch of the left coronary artery. The patient's postoperative course was uneventful, and she was discharged on postoperative day 15.

Discussion

A coronary-pulmonary artery fistula is an occasionally observed congenital anomaly, but it is rarely associated with a coronary artery aneurysm. It causes few symptoms and is often detected incidentally with a reported incidence of 0.3%–0.8% among all coronary angiographies.[1,2] It has been reported that coronary-pulmonary artery fistulas account for 15%–20% of all coronary artery fistulas and are associated with aneurysmal dilatation in 19%–26% of cases.[3] The left anterior descending branch, both the left anterior descending branch and right coronary artery, the right coronary artery, both the

Fig. 3. Postoperative coronary angiography showing the disappearance of the coronary artery aneurysm and the abnormal blood vessels.

left anterior descending and left circumflex branches, and the flow into coronary artery aneurysms in percentage of cases are as follows: left anterior descending branch (55%); left anterior descending branch and right coronary artery (26%); right coronary artery (8%); left anterior descending branch and left circumflex branches (6%); other arteries (5%).[4] Also, 16%, 35%, 16%, and 10% of fistulas have been reported to open into the right ventricle, right atrium and coronary sinus, pulmonary artery, and left side of the heart, respectively.[5] Although congenital coronary-pulmonary artery fistulas and coronary arteriovenous fistulas are known to become aneurysmal, these aneurysms are mostly fusiform and rarely saccular. The causes of coronary artery fistulas complicated by saccular coronary artery aneurysms include turbulent flow and arteriosclerotic changes as a result of acquired inflammation, trauma, kinking, and stenosis; in particular, those of atherosclerotic origin are the most common.[6] In this patient, the histopathological findings showed that the aneurysm was due to atherosclerotic changes with fibrosis.

Most patients with this disorder are asymptomatic, but some patients present with angina-like chest pain, exertional respiratory discomfort, shortness of breath, and palpitation, presumably because of a shunt-induced decrease in coronary blood flow. Some patients, including this one, present with a continuous heart murmur. With advances in multidetector row CT (MDCT), it has become possible to preoperatively determine the morphology of coronary artery aneurysms and the course of abnormal arteries as sharp images. However, coronary artery aneurysms are frequently accompanied by stenotic lesions, and their accurate diagnoses requires concomitant coronary angiography. Because of the risk of rupture and coronary embolism, surgery is often necessary. Some studies have reported that a coronary artery aneurysm more than 3 cm in diameter is an absolute indication for surgery.[7,8] On the other hand, even small saccular aneurysms complicating coronary-pulmonary artery fistulas have been reported to rupture, suggesting that surgery should be considered even in the absence of symptoms. The risk of rupture cannot be determined, at least in terms of aneurysm diameter, and many opinions suggest that surgery be performed proactively in patients with coronary-pulmonary artery fistulas complicated by coronary artery aneurysms. Although the present patient had no evidence of myocardial ischemia and was asymptomatic, we decided to perform surgery because the 3-cm-diameter aneurysm carried the risk of rupture. According to Konno and Endo,[9] the surgical indications for this condition are (1) a shunt of more than 30%; (2) electrocardiographic evidence of ischemic changes; (3) advanced pulmonary hypertension; (4) advanced congestive heart failure; (5) a history of infectious endocarditis; (6) the risk of rupture resulting from aneurysmal change; and (7) the social influence of heart murmurs. Coronary-pulmonary artery fistulas used to be treated by ligation without extracorporeal circulation. In recent years, besides the ligation of fistulous vessels under extracorporeal circulation, the fistula is directly closed from the inside of the pulmonary artery, depending on the patient's condition, thereby enabling the prevention of fistula persistence after surgery. Coronary-pulmonary artery fistulas that have undergone aneurysmal change require the closure of blood vessels flowing into the aneurysm and the resection or suture reduction of the aneurysm. Some patients need concomitant coronary artery bypass surgery, depending on the presence or absence of coronary artery stenotic lesions and the location of coronary artery aneurysms.[10] Because the patency of the vascular openings in the aneurysm was confirmed in this patient, we performed direct suture closure of the inflow vessels, direct closure of these openings from inside the aneurysm, and aneurysmorrhaphy, all with good results. There was no need for coronary artery bypass surgery.

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